

CYANOSIS RETINÆ.¹

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THIS term is used to describe the changes in the fundus which are observed in congenital heart disease with general cyanosis, and was first employed by Liebreich, in 1863, in his report of a case of this kind, the illustration of which may be found in the first edition of his *Atlas*. The honor of being the first to call attention to the changes in the eye-grounds, designated by the term of "cyanosis retinæ," however, belongs to Knapp, who exhibited a water-color and described the ophthalmoscopic findings in such a case before the Heidelberg Society in 1861. Von Ammon² had given a short description and sketch of the exterior of the eye in congenital heart disease in 1841, but no study of the fundus had been made, as the ophthalmoscope had not yet been discovered.

Since the publication of Knapp's description other cases have been recorded, although their number is exceedingly small, in view of the comparative frequency of congenital disease of the heart. This is to be accounted for, in all probability, not by the rarity with which this form of heart disease causes changes in the retina, but rather by the failure of clinicians to examine the eye-grounds in these cases, or, in the event of such examinations being made, to allot any special significance to them.

As may be seen by the illustration, from a case recently observed by the writer, and the description of which will follow, the changes revealed by the ophthalmoscope in cyanosis retinæ are most striking, the vessels of the retina attaining a size which is rarely equalled in any other condition, this being particularly true of the veins, which may become enormously dilated and tortuous; the arteries, though usually similarly distended, being affected to a less degree. In addition to the change in their calibre, both veins and arteries become much darker than normal, the former assuming a deep-violet color, while the arteries resemble normal veins. The peripheral twigs of the retinal vessels are distended, and vessels which are usually invisible may be seen over the entire fundus. Pulsation in the vessels is absent. Small hemorrhages are of frequent occurrence in the neighborhood of the disk; more rarely larger ones are found in the macula, when vision may be much affected.

¹ Read before the American Ophthalmological Society, May, 1905.

² Clinical Representation of the Diseases and Malformations of the Human Eye (Tafel, vol. xv. Fig. 3, Text, Bd. iii. p. 75.

As is well depicted in von Ammon's sketch, the exterior of such eyes also present marked signs of cyanosis, the lids becoming blue, the sclera violaceous, while the vessels of the conjunctiva are much distended. In the later editions of his *Atlas*, Liebreich withdrew the illustration of the case of cyanosis retinæ which had appeared in the first edition, and substituted for it one of abnormal pigmentation of the eye, to which he inappropriately applied the former name. Although, owing to the popularity of the *Atlas*, this change occasioned for a time some confusion regarding the precise application of the term, the subject is now well understood, and, as indicated in the opening sentence of this paper, the name of cyanosis retinæ is applied to the eye changes observed in cases of congenital heart disease alone.

The congenital diseases and malformations of the heart which may occasion changes in the fundi are various. Thus, they may be found where there is an abnormal connection between the right and left ventricles, and in cases in which there is a stenosis of the pulmonary artery or an abnormal origin of the aorta and pulmonary artery. They may also arise when there is a general retardation of the blood current, as a consequence of congenital dilatation of the entire vascular system.

The cause of the dilatation of the retinal vessels has been a matter of some dispute among clinicians, some maintaining that it is simply the result of the general venous stasis, while others aver that this cannot be true, as retinal changes rarely, if ever, occur in cases of general cyanosis due to causes other than congenital heart disease, and contend, moreover, that the arteries often manifest changes as well as the veins. Most authors attribute the dilatation of the vessels to a stretching in the walls of the vessels themselves, and consider this to be dependent upon a lessened resistance in the walls, due to disturbances in nutrition occasioned by insufficient oxygenation of the blood.

Although the examination of the eye-grounds in cases of congenital heart disease has not been considered by clinicians to be of particular moment, the dilatation and tortuosity of the retinal vessels being regarded as merely additional evidence of the general cyanosis, recent observations by Babinski and Nagel indicate that the ophthalmoscope may be of decided value both in the diagnosis and prophylaxis of this class of heart cases. Thus, Babinski has reported a case in which a marked dilatation of the retinal vessels preceded other evidence of general cyanosis, while Nagel, from a study of the cases in the literature, and from his own observation, asserts that if the ophthalmoscopic examination reveals that the retinal arteries as well as the veins are darker and broader than normal, the presence of a connection between the two sides of the heart is assured; when the darker coloration and tortuosity is limited to the veins, he believes that the existence of some other

congenital lesion is indicated, which does not permit the admixture of venous with arterial blood.

If these observations are true, clinicians should be made fully cognizant of them, for it is generally conceded that the diagnosis in cases of congenital heart disease is at times practically impossible from a study of the physical signs. It is hoped, therefore, that the practice of examining the eye-grounds in this class of cases may become more universal, and that the results of such examination be recorded, as it is only by means of a series of such observations that the assertions of Babinski and Nagel can be confirmed, and the actual value of the ophthalmoscope to the clinician proven.

The vision in eyes the seat of cyanosis retinæ may be normal, but if hemorrhages occur, as is not infrequently the case, vision may be more or less involved. Both eyes are usually similarly affected.

The attention of this Society was first called to the subject of cyanosis retinæ by Knapp, in 1870, who exhibited before the annual meeting of that year the sketch which he had prepared of the case exhibited by him in Heidelberg, and gave a short *résumé* of his notes. Since that time no further mention of the condition has been made before the Society, so that the writer considered that a brief *résumé* of the cases which had already been reported, and the details of two additional cases, might be not without interest.

1. As already recorded in the *Transactions* of the Society, in Knapp's case there was non-inflammatory, excessive hyperæmia of both retinæ, occurring in a patient with enlargement of the heart, but without valvular anomalies, who exhibited general cyanosis and aneurysmal bruit at different parts of the body. The retinæ were so vascular that a countless number of thick and tortuous arteries and veins arose from the optic disks and branched radiating toward the periphery, like a caput Medusæ. The area and borders of the optic disks were totally concealed. The yellow spot was distinctly discernible. A great number of arterial and venous twigs reached to the fovea centralis. The retina, refracting media, sight, and visual field of both eyes were perfectly normal. The patient died in the hospital at Heidelberg. The body was found to exhibit a general enlargement of the whole vascular system.

2. The next case was observed by Liebreich, in 1863, and, as already stated, was illustrated in the first edition of his *Atlas*. Examination of the vascular system showed stenosis of the pulmonary artery, with considerable hypertrophy of the right ventricle. There was general cyanosis. The ophthalmoscope showed the retinal veins to be twice as broad as normal, without being tortuous or presenting unevenness. The arteries were normal.

3. Leber¹ reported the third case, calling particular attention

¹ First edition of the Graefe-Saemisch Handbuch, Bd. v. p. 525.

to the color of the blood, which was unusually dark in the arteries, being about the same color that the veins normally present, while in the veins it was dark violet-brown. The vessels were full and stood out from the level of the eye-ground. The anterior ciliary veins were unusually dark and the conjunctiva cyanotic. The patient was the subject of general cyanosis, and was thought to have stenosis of the pulmonary artery. This author, in his capacity as assistant, had had an opportunity of studying Knapp's case, and referred to it at length in his treatise in the first edition of the Graefe-Saemisch *Handbuch*.

4. In 1878 Stangloneier¹ reported a case of pulmonary stenosis in which there was endocarditis of the pulmonary valves, a patulous foramen ovale and ductus Botalli, small retinal arteries and very tortuous veins, with two large hemorrhages in the nasal part of the retina, which appeared shortly before death.

5. In the next case, a man aged twenty-five years, reported by Litten² in 1882, the fundus was found to be abnormally dark, emitting a decidedly bluish reflex. The retinal veins also were enlarged and tortuous. Examination showed a complicated valvular defect in the ostium pulmonale, associated with a defect in the interventricular septum. The same author³ referred to another kind of cyanosis retinæ, namely, that produced by aniline poisoning, and cited a case in which the conjunctiva and fundus appeared violet and the retinal bloodvessels almost black. Vision was unaffected.

6. Nagel reported the sixth case, occurring in a boy aged nine years. One eye was staphylomatous, following a perforating ulcer of the cornea three years previously. The fundus of the fellow-eye showed the typical changes of cyanosis retinæ, the entire eye-ground being dark red and the veins almost black. The author was of the opinion that the marked cyanosis, the dyspnœa, the coolness of the skin, the clubbing of the fingers, pointed to a stenosis of the pulmonary artery, with probable mitral insufficiency.

7. In his monograph on *Congenital Affections of the Heart* (p. 23) George Carpenter cited a case which occurred in a child five and a half years old. There was extreme general cyanosis, the face and body being dusky, the mucous membrane slate-colored and the conjunctiva suffused. Ophthalmoscopic examination "showed the retinal veins to be *very* large, the arteries of good size, and both sets of vessels most intensely cork-screwed. There was no edge to the optic disk, and the red reflex apparently started from the physiological pit." At the autopsy both ventricles were hypertrophied and dilated, the right being in excess. The right auricle was twice

¹ Inaug. Dissert., Würzburg, 1878; Nagel's Jahrbuch f. Ophth., 1878, p. 261.

² Deutsche med. Wochen., 1887, No. 8, p. 144.

³ Berl. med. Wochen., 1881, Nos. 1 and 2.

the thickness of the left, but the left was dilated and the right auricular appendix was four times the size of its fellow, which was decidedly small. There was a patent septum ventriculorum, capable of admitting the index finger. The foramen ovale was closed. The aorta arose equally from both ventricles, and there were further abnormalities in its branches.

8. The same author referred to a second case, seen by him, in a small boy who was somewhat cyanosed. No cardiac bruit was detected. An ophthalmological examination revealed fundal changes similar to those noted in the case just reported, and suggested to him the nature of the complaint.

At the close of his interesting monograph Carpenter gave the notes of 22 cases of congenital heart disease, but unfortunately an ophthalmological examination appears to have been made in but one instance, where the history contains the note that "the veins of the optic disk pulsate and appear beaded."

9. In Loring's *Text-book of Ophthalmology* (vol ii. p. 106) will be found the notes of an additional case, occurring in a young woman aged twenty-eight years. Loring reports: "She had never noticed that her skin was darker than that of other people until six years ago, when she was shipwrecked, during which she was much terrified. Her finger-nails were, however, club-shaped, and she had been informed by her mother that this had been the case since she was ten months old. Her skin is now of a livid, bluish tinge, with dark-purple spots upon the face and hands, and she thinks that the color is increasing in darkness. The ears are purplish and the conjunctiva a good deal injected.

The veins of the fundus are enormously enlarged and tortuous, and have an anteroposterior curve in certain places, but only on the disk and its immediate neighborhood. There are very many vessels that are plainly visible and can be traced far into the retina, which, in the normal state, are either not present or are so minute as to be invisible. The color of the blood in the veins is of a violet tinge. The arteries are considerably enlarged, but not so much as the veins. The blood in the arteries is of the color of ordinary venous blood. There is no pulsation in either arteries or veins. An exhaustive examination by an expert failed to bring to light any disease of the heart or any abnormality in its sounds. The ophthalmoscopic picture presented a strong similarity to that of choked disk, and had been, indeed, taken for that."

10 and 11. In 1892 Hirschberg observed two cases. In the first the retinal veins were dark blue, tortuous, and dilated. There were, in addition, general cyanosis, fingers clubbed, pulmonary emphysema, dilatation of right heart, and vision reduced to one-quarter in each eye. In the second case the heart was malformed and there was the same appearance of the eye as in the preceding case. Vision was normal.

12. Another case was reported by Galezowski,¹ in which vision was said to have been much lowered by intense congestion of the retina in the macular region, and, as in Stangloneier's case, retinal hemorrhages appeared a few days before death.

13. In 1899 Fehr reported a case before the Berliner Ophthalmologische Gesellschaft, April 20, 1899, but the writer has not had access to the *Transactions* of the Society, and has been unable to find the notes of the case elsewhere.

14. Quite recently Goldzieher² reported an interesting case of congenital disease of the heart with hyperglobulia, in which the left eye was lost, as a consequence of hemorrhagic iridocyclitis, and the right eye presented the typical changes described as cyanosis retinæ. The patient was a female aged nine years, who had always been sickly and had suffered repeatedly from nasal hemorrhages. The exterior of both eyes presented marked signs of cyanosis. The retinal arteries and veins of the right eye were much enlarged and darkened, and the edges of the disk were everywhere sharply outlined; there were no hemorrhages. The iris of the left eye, however, was stippled with small hemorrhages, and the vitreous contained so much blood that no reflex could be obtained from it. The globe was stony hard, and finally underwent spontaneous rupture through the sclera at the equator. Dr. Hochwall, who made a careful study of the case clinically, found a considerable increase in the hæmoglobin in the blood, as a consequence of increase in the number of red blood corpuscles, which he considered to be an expression of increased concentration of the blood. He thought the hyperglobulia was dependent upon the disturbance in the circulation, as a consequence of the congenital affection of the heart, and was not an instance of the symptom-complex in which there is an association of polycythæmia, cyanosis, and tumor of the spleen. The determination of the precise nature of the cardiac lesion was difficult, but the physical signs and the employment of the x-rays indicated that either the foramen ovale was patulous or that there was a defect in the septum atrium.

15. In 1904 Babinski³ saw a case in a ten-year-old subject with stenosis of the pulmonary artery. General cyanosis first appeared at three years of age, during an attack of whooping-cough. Left-sided hemiplegia developed at this time also. The retinal arteries were tortuous and dilated, and the veins were enormously enlarged.

16. In a recent number of the *Annales d'Oculistique*, February, 1905, the same author, in conjunction with Toufesco, reported another case occurring in a male aged thirty-five years. The patient had been sickly, but symptoms of cardiac congestion first became manifest after a prolonged attack of typhoid fever, when

¹ No reference. Cited by Babinski, *Revue neurologie*, 1904, vol. xii. p. 1146.

² *Centralblatt f. prak. Augenheilk.*, September, 1904.

³ *Revue neurologie*, 1904, vol. xii. p. 1143.

seventeen years of age. The pulmonary artery was stenosed. *Cyanosis of the retina was found without general cyanosis, both arteries and veins being markedly dilated.* The authors conclude from this that general cyanosis as a consequence of stenosis of the pulmonary artery may manifest itself in the retina before changes appear in the skin.

17. In 1903 a case of cyanosis retinæ was observed by the writer, which had been referred to him at his service in the Howard Hospital by Dr. S. M. Hamill, who had made a careful study of the general condition of the patient, which he later embodied in a report which has been published elsewhere.¹ The abbreviated history was as follows: The patient, a boy aged nine years, first became blue when a few months old. Dyspnoea had always been marked upon exertion. At the time of examination the boy was noted as being of normal size for his age and well nourished. The skin of his face and body and all the visible mucous membranes had a deep, purplish color. The skin was rough and dry and was covered with small, bluish follicles. The hands and feet were especially congested, and clubbing of both fingers and toes was present to a marked degree. Examination of the heart showed a double impulse, which was most marked in the fourth interspace. The left chest was more prominent than the right, but there was no thrill. At the apex there was a rather soft, short, blowing, early systolic murmur, which accompanied the first part of a divided first sound. The murmur and the divided sound were both heard in the axilla, over the body of the heart, and best in the third space to the left of the sternum. The murmur was heard faintly in the aortic area. The second pulmonic sound was much accentuated. The heart's action was fairly regular and rapid. There were marked pulsations in the veins of the neck and in the subclavians. Posterior to the sternocleidomastoid muscle of the left side, and almost directly underneath the mastoid process, there was a marked pulsation, which on palpation felt like a tortuous and anomalously located external carotid artery. Dr. Hamill was unable, from the physical examination, to reach any satisfactory conclusion regarding its specific character, but inclined to the view that the case was one of stenosis of the pulmonary artery, with patulous ductus arteriosus, basing this opinion upon the age of the patient, the location of the murmur, and the greater frequency of this form of cardiac lesion.

As incorporated in Dr. Hamill's report, the condition of the eyes was as follows: The eyeballs were prominent. The conjunctival vessels, both those of the globe and those of the lids, were swollen, especially the veins, giving the eye a violaceous appearance. The pupils were large and the media clear. Vision was 5/6 in each eye.

¹ Pediatrics, May, 1903.

The ophthalmoscope showed a neuroretinitis of marked degree. The retinal arteries and veins were greatly swollen and tortuous, resembling large angle-worms. The writer does not remember ever to have seen retinal vessels of a larger calibre, the retinal veins appearing to be increased to three times their normal size. The head of the optic nerve was obscured by the swollen retina, and there were a few small hemorrhages into the nerve-fibre layer of the retina close to the disk.

18. About the same time that Dr. Hamill's case was under observation, Dr. W. C. Swindells, the office assistant of the writer, found similar changes in the eye-grounds in a case of Dr. Howard Fussell, to whom he is indebted for the following notes: "Male child, aged one year; presented no abnormality during the first two weeks of life. When next seen by Dr. Fussell, however, about a year later, marked signs of cardiac disease had manifested themselves. It was elicited from the mother that when the child was about two or three months old, and while he was lying in the cradle, he was suddenly seized by an attack of unconsciousness, during which his entire body became blue. As he grew older and moved about, attacks of dyspnoea became frequent, during which the face and fingers became blue. Occasionally, and especially when he was disturbed or attempted to run, attacks of unconsciousness supervened, during which the finger-nails became blue and the lips cyanosed. Examination revealed that the fingers were clubbed; the precordia was prominent, and there was a marked prominence over the junction of the first and second piece of the sternum. The heart dulness was at the right border of the sternum and first rib diagonally downward to the nipple line. There was a very rough systolic murmur heard all over the chest, with greatest intensity at the upper border of the heart dulness, and transmitted into the vessels of the neck.

Shortly after this, and while still under Dr. Fussell's observation, the child had an attack of what was considered to be pneumonia and later was ill with true diphtheria with laryngeal stenosis. During the treatment of the diphtheria 2000 units of antitoxin were administered, with no untoward effects. After convalescence it was noted that the heart dulness had moved still farther to the right and was broader at the apex. A month later the cyanosis was more marked and the dyspnoea had become constant. The heart's action was slow and regular. A loud, high-pitched systolic murmur was heard over the aortic cartilage. The second heart sound was accentuated. To the left of the sternum the murmur was not as loud nor the second sound as sharp. The murmur was heard, however, over all the anterior part of the chest, its point of greatest intensity being just over the sternum, below the junction of the first and second parts. It could be faintly heard in the vessels of the neck and in the back. There was no pulsation in the

external jugular. The second sound was loudest midway between the nipple and sternum, over the fourth interspace. The edge of the liver could not be plainly felt, but there was more resistance on the right side than on the left. The upper line of the liver dulness corresponded to the fifth rib in the axilla. There was no œdema of the legs. The child gr̄ew gradually worse and died a few months later with symptoms of cardiac failure. No autopsy was permitted; so that the probable diagnosis of mitral regurgitation, with, perhaps, some communication between the two sides of the heart, could not be ratified.

Owing to the fact that the diagnosis of the cardiac condition in the 18 cases which have been just cited was confirmed by autopsy in but four instances, and that the descriptions of the changes in the eye-grounds in most cases are too meagre from which to draw inferences regarding the precise lesion in the fundus which the various cardiac lesions may occasion, it has not seemed worth while to attempt to summarize the facts which have been given, or to seek to prove the truth of Nagel's assertion from them. As indicating the value of ophthalmoscopic examination in cardiac cases of this type, it is of interest, however, in addition to the case of Babinski, which has already been cited, to refer to Carpenter's second case in which there was no cardiac bruit and but slight cyanosis, and the true nature of the complaint was determined only after the discovery of the typical changes of cyanosis retinæ by the ophthalmoscope.

SCARLATINAL OTITIS.

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STRANGE as it may seem in these days of advanced thought in medical science, there are yet a few physicians who are firmly adherent to the old notions of tentative medicine.

After all that experience has taught as to the dangers to the functions of hearing and of life itself by ear inflammation complicating scarlet fever, yet there are many of our profession who assume an attitude of expectancy and contented helplessness, and a peculiar sense of satisfaction when an ear aches, breaks, and runs during the course of this infectious disease.

This is unfortunate for the science of medicine and for the human race, and inexcusable in these days of enlightenment.

We might excuse the doctor of past generations, who felt that scarlet fever must leave some "corruption" in the system and felt